MEDICAL PRACTICE

Occasional Review

Prognosis of the Ramsay Hunt syndrome

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Summary and conclusions

Thirty-six cases of herpes zoster complicated by facial paralysis (the Ramsay Hunt syndrome) seen over 10 years were reviewed to determine the subjective degree of recovery (ascertained by questionnaire) and residual disability (ascertained by re-examination). Eighteen patients made a full recovery, most within three months; 14 patients were left with only mild residual signs. In only four patients was the final result regarded as unsatisfactory. Outcome was not adversely influenced by age.

The facial paralysis of the Ramsay Hunt syndrome thus carries a generally favourable prognosis.

Introduction

The Ramsay Hunt syndrome of geniculate herpes, which Ramsay Hunt himself ascribed to inflammation of the geniculate ganglion, consists of herpes zoster of the auricle, with facial paralysis and loss of taste. A herpetic rash may also appear in the trigeminal area, or in an area corresponding to the distribution of the glossopharyngeal nerve, and sometimes in the upper cervical region. The eighth, sixth, and lower cranial nerves may also be affected, with deafness or vertigo, diplopia, and dysphagia, respectively. In severer cases there may be fever, meningism, and mental confusion.[1-3]

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Standard textbooks of neurology suggest that the prognosis for recovery from the facial paralysis is poor,[4 5] although Taverner thought that it was as good as in Bell's palsy.[6] Previous studies, however, have included only small numbers of patients.[7-9] We have therefore reviewed all the cases seen by one of us over 10 years in an attempt to obtain a more accurate idea of the outcome.

Patients and methods

The series consisted of 36 patients, 17 men and 19 women, with geniculate zoster seen between 1967 and 1976. Their age distribution, characteristic of herpes zoster in general, was as follows: 21-30 years, 2 patients; 31-40, 1; 41-50, 3; 51-60, 16; 61-70, 5; and 71-80, 9,

A questionnaire was sent to all those not under current review, asking for information about the degree of recovery and any additional observations; those patients reporting residual facial weakness were then given a further appointment.

Electromyography was carried out only on those patients who were not recovering from the paralysis within a month. We did not routinely look for antibodies to herpes zoster virus as all the patients had typical herpetic eruptions. The cerebrospinal fluid was examined only in those patients with possible meningoencephalitis. Corticosteroids were given in selected cases.

From the combined results of the questionnaires and clinical assessment recovery was graded as nil, minimal, moderate, good but with some residual disability, and complete. The length of follow-up until patients were discharged from the clinic was also used as a marker of recovery.

Results

In all patients the facial paralysis had been complete when they were first seen. The right and left sides were affected in equal numbers. In most the rash had appeared before the paralysis, but in two a week after it and in two at the same time. One had had herpes zoster in the thoracic region 18 years previously.

The rash most commonly occurred on the ear, pinna, and auricle (16 patients; table I), and it followed the distribution of more than one

TABLE I—*Site of rash in 36 patients with geniculate herpes*

Site	No of patients
Distribution of trigeminal V_1	3
V_2 face	7
V_2 hard palate	5
V_3	7
Soft palate	1
Eardrum	1
Ear (including pinna and auricle)	16
Neck	3*

*In one the rash also included the upper chest in the distribution of the fourth cervical nerve.

nerve in eight patients; in no case was it confined to the sensory distribution of the facial nerve.[10] A few patients had eruptions on the neck, soft palate, and eardrum—in the last case it was the only cutaneous manifestation; the lesion might easily have been missed and Bell's palsy diagnosed. Unlike patients with herpes zoster in general,[11] the first division of the nerve was affected in only three of the 22 patients whose rash had a trigeminal distribution.

RECOVERY

All 36 patients were followed up until they had achieved their maximum recovery; this took from under three months to more than a year, 21 being discharged within six months and 12 within three months (table II). Eighteen made a full recovery, 14 a good recovery but with mild residual facial weakness, three a moderate recovery, and one no recovery until after a faciohypoglossal anastomosis. The three patients with moderate recovery required tarsorrhaphy, one permanently.

TABLE II—*Degree of recovery and time for maximum improvement in 36 patients with geniculate herpes*

Degree of recovery	No of patients	Months to discharge	No of patients
Nil	1	<3	12
Minimal	0	3–6	9
Moderate	3	6–12	10
Good but with residual disability	14	>12	4
Full	18		

COMPLICATIONS

Sixteen patients had no neurological signs other than a rash and facial paralysis. The commonest complications (in 17 patients; table III) were due to lesions of the vestibular branch of the eighth nerve —namely, mild labyrinthine ataxia in seven patients, with veering towards the affected side but no nystagmus; and vertigo in 10, five of whom had nystagmus.

Fourteen patients had some deafness. In most this was due to exudate in the external auditory meatus, but in two patients it proved to be permanent nerve deafness.

Nine patients (eight of them with ataxia or deafness) complained of loss of taste; this was transient in all except one. Sixth nerve palsies occurred in two patients with diplopia, lasting two and three weeks. Only two patients developed postherpetic neuralgia.

Lumbar puncture—Three of the four patients admitted to hospital had lumbar punctures. In two the cerebrospinal fluid was abnormal, containing 120 and 1800 lymphocytes per litre, the CSF protein in the latter case being raised to 1·48 g/l (148 mg/100ml).

Electromyography—Electromyograms done in eight patients showed that latencies were normal in two of the three patients tested, and these made an excellent recovery; whereas the patient with a prolonged latency had residual contracture of the facial muscles after recovery. Fibrillation potentials were present in the other patients tested.

Steroid treatment—Eight patients were given steroids—five intramuscular ACTH, 40 units daily; and three prednisolone, in an initial dose of 80 mg daily. Two of the patients given ACTH did well, but a third appeared to derive no benefit; of the remaining two, one did not receive an adequate course of treatment while the other did not start the drug until three weeks after the onset of paralysis. In the three patients given prednisolone recovery ranged from moderate to full.

TABLE III—*Complications of geniculate herpes in 36 patients*

	No of patients
Loss of taste	9*
Vestibular involvement	17
ataxia	7
vertigo	10
nystagmus	5
Deafness	14†
Sixth nerve palsy	2
Postherpetic neuralgia	2
Varicella	1

*Permanent in one.
†Permanent in two.

Discussion

In his original review of 60 cases of herpes zoster associated with facial paralysis Ramsay Hunt found that complete facial palsy could either clear rapidly "lasting only a few days or a fortnight" or be prolonged and lead to "permanent weakness and contractures of the face,"[1] though he did not give exact figures. Our series of 36 patients suggests that few patients with geniculate herpes will be left with much disability: half made a complete recovery, and only four had an unsatisfactory degree of recovery. These four patients were all in their 50s, and as no fewer than nine were over 70 in this series age would not appear to give an adverse prognosis. Over half the patients were discharged from follow-up within six months, and a third within three months.

The complications were often transient. Vestibular symptoms were the commonest, and were mild and transient in 14 patients; vertigo invariably cleared in a few weeks. Only two patients had permanent hearing loss.

As standard textbooks suggest a gloomy prognosis we think that it is important that the largely favourable outcome in our series should be more widely known, so that patients can be reassured about their distressing and sometimes bizarre symptoms. The value of steroid treatment, however, can be assessed only if the drug is given promptly and in adequate dosage and the results are compared with those of conservative treatment in similar numbers of patients.

References

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ONE HUNDRED YEARS AGO An application was made last week, in the County Magistrates' Court, Liverpool, by Inspector Johnston, for an order for the confinement of all dogs within the Kirkdale division of the county, owing to the prevalence of hydrophobia. He instanced several cases of hydrophobia arising from dog-bites recently. An order was granted, to be in force to the end of February. Another death has occurred from hydrophobia near Taporley, Cheshire, making the third in the same district within the space of three weeks. (*British Medical Journal*, 1878.)